

Diffuse Large B Cell Lymphoma Mimicking a Large Soft Tissue Sarcoma of the Shoulder

Case report in a 42 years old man, follow-up and review of literature



Healthcare

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Abstract

There are different complex anatomical structures in the shoulder and axillary regions, many of which can give rise to neoplasms. Lymphomas with the involvement of soft tissues as a primary event are very rare. The published studies have a small sample size, most of them as case reports. We want to highlight and draw attention to the clinicians, surgeons, pathologists and all the doctors to pick up this pathology in the differential diagnosis in a tumor which is first suspected to be a soft tissue sarcoma. We report a rare case of shoulder pain in a patient affected by a Diffuse Large B-Cell Lymphoma (DLBCL), of the left shoulder, his follow-up and a review literature of the shoulder lymphoma.

Introduction

Even though most shoulder affections are diagnosed by history, physical examination, and radiographic imaging, diffuse neoplasms of this area may be better assessed at histopathology(1). Among these latter, the non-Hodgkin lymphoma (NHL) is one of the most common, especially the diffuse large B cell lymphoma (DLBCL). The incidence of NHL is increasing, accounting for 40,000 cases per year in the United States(2), mostly in patients with acquired immunodeficiency or taking immunosuppressive drugs. Often lethal, it usually occurs with multiple lymphadenopathies, but any site containing lymphoid tissue, included the bone marrow, may be involved. The involvement of soft tissues is primary in less than 1%(3) and, when present, is usually secondary to a direct spreading from affected surrounding lymphoidal sites and/or metastatic hematogenous dissemination (4). From a clinical point of view, lymphomas usually manifest with a soft tissue mass, swelling, and pain. Most patients are asymptomatic whereas the main presentation is a painless adenopathy in the neck or supraclavicular region (5).

Soft tissue sarcomas and lymphomas are among the malignant neoplasms which can be seen in these regions. Differentiation between these tumour types is based on clinical findings, radiological features, and especially in histopathologic examination. The histopathological distinction between lymphoma and soft tissue sarcoma is routinely determined by IHC. There is a large panel of immunohistochemical markers like: Leucocyte common antigen (LCA) (lymphoid marker), CD3, CD20 and vimentin (mesenchymal marker), which are commonly used for this purpose.

Case Report

In August 2013, in our hospital, a 42-years old man presented with diffuse left shoulder mass, it has been present for two years with progressive enlargement, which was associated with continuous pain and difficulty in the movement of the left scapulo-humeral articulation. He denied any constitutional symptoms, including weight loss, fever, or night sweats. On physical examination the mass was poorly defined with a smooth outer surface, and according to these data, clinically a sarcoma of soft tissue was suspected. Clinico-biochemical analysis was in norme also tumoral markers. (HCG<2mIU/ml, AFP-0, 6UI/ml, CEA-0,5 µg/L). CT-scan and MRI revealed a diffuse infiltrative tumour mass involving left shoulder musculatures. Figure 1(a,b,c,d).



Figure 1a

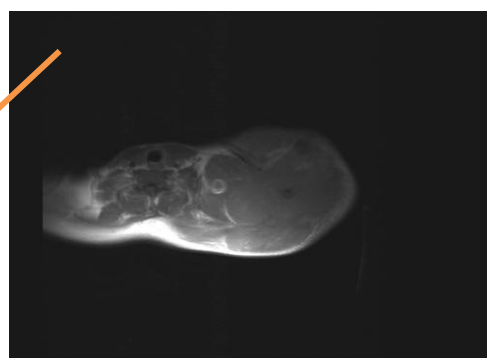


Figure 1b

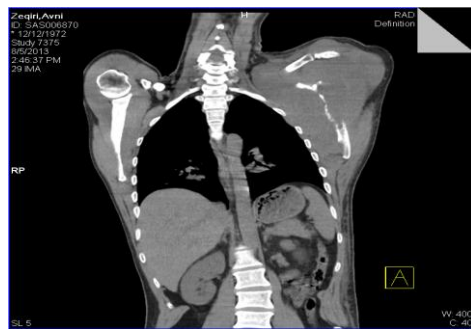


Figure 1c

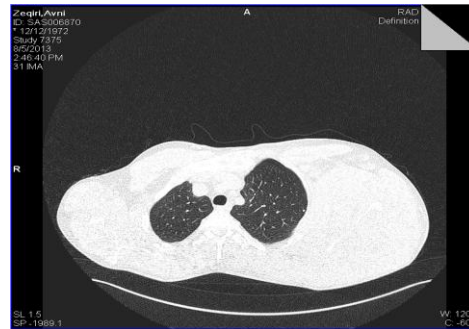


Figure 1d

Figure 1(a,b,c,d)-MRI and CT scan show a diffuse soft tissue mass involving the left shoulder musculatures, but without involvement of left lung.

In this condition, with the consent of the patient an incisional biopsy of the mass was performed. Microscopic examination showed diffuse cell infiltration with big round nucleus, with evident nucleolus and frequent mitosis. Immunohistochemistry performed showed: CD45(+), **CD20(+)**, CD79a(+), CD3(-), CD23(-), CD30(-), Bc-2(-), Bcl-6(-), Bcl-1(-), TdT(-), Vimentine(-) and Ki-67 65-70%, which are consistent with: **Diffuse large B cell lymphoma**.

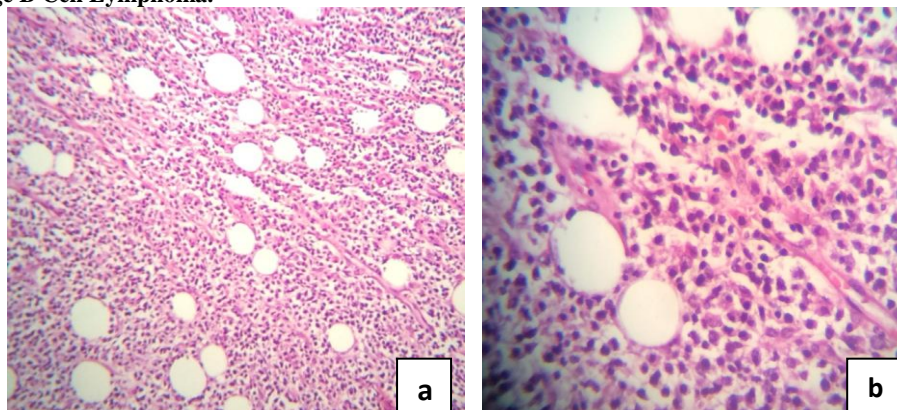
The patient has done 9 cycles of chemotherapy (CHOP+Rituximab) after 8 months (April 2014) he comes again to the hospital with a huge mass in the same location. The patient has limitation of movement in scapulohumeral articulation, large pain, edema and echimotic changes of the skin above the tumor as in (the figure 2) below:



Figure 2: After second en bloc resection of the tumor surgery with some large diffuse soft masses, where the biggest one is 15*10 cm.

Immunohistochemistry of paraffin sections showed positive membranous immunoreactivity in CD79a(+), Bcl-6(+), Bcl-2(-), CD3(+), CD5(+), CD10(-), CD23(-), Vimentine(-) in stromal cells, but negative in neoplastic cells, which exclude mesenchymal histogenesis (Figure 3).

Most important findings was **CD20(-)**, which is connected with ANTI-CD20-Therapy. So these findings were consistent with **Diffuse Large B Cell Lymphoma**.



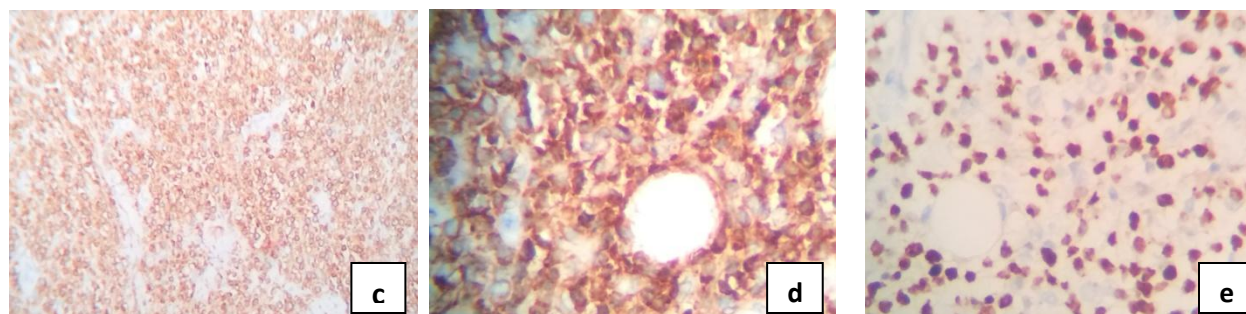


Figure 3 Hematoxylin and eosin (a, b) staining of the lesion and some immunostaining images (c-e).
Shown are immunostaining examples of CD79a (c), CD20 (d), Ki-67(e).

He underwent again chemotherapy 5 cycle. In September 2014 he complains physical weakness, fatigue, pale mucosae and skin, thoracic discomfort, anorexia, and bleeding from the mass. The inspection of the scapulohumeral articulation revealed laceration and destruction of the skin and all surrounding soft tissues around scapulohumeral articulation and scapula, accompanied with hemorrhagic changes, as in (the figure 4) below:

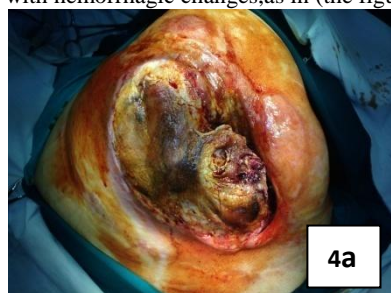


Figure 4 Fig.4a before operation



Fig.4b Specimen

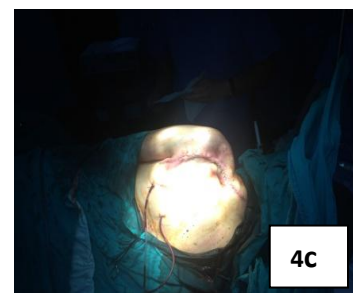


Fig.4c after operation

Second surgery was performed where is observed including of glenoidal capsula. To date the patient is following chemotherapy and his physical condition is not so good.

Discussion

DLBCL is a group of large, lymphoid B-cell malignant proliferations that is clinically, morphologically, and genetically heterogeneous. It constitutes about 30% of all non-Hodgkin lymphomas and is the most common histologic subtype (6). A shoulder lymphoma has to be differentiated from neoplasms involving the soft tissue, mostly the sarcoma, osteomyelitis, spine affections, and neurological disorders. The case reported here was initially considered rhabdomyosarcoma, because of the morphologic features, the patient's age and the location of the tumor. MRI is more specific to assess the extent of the mass within the bone and surrounding soft tissues. The initial diagnosis was ruled out by immunohistochemical analysis that showed that tumor cells were positive for CD20, CD45 and negative for vimentin. This is the primary case of Diffuse Large B cell-lymphoma of the shoulder occurring with isolated pain and limited range of motion of the left shoulder in an Albanian patient without any associated immunological disorder. Lymphoma may also be associated with lupus erythematosus (7), scleroderma, and Sjögren's syndrome. In addition, prolonged immunosuppressive therapies may increase the risk for neoplasms of the immune system (8).

The malignancy's stage and histopathological diagnosis have a major impact on its treatment and prognosis. Patients with lymphoma are usually treated with chemotherapy or local irradiation. Up to now, there are a few cases reported in the literature. So, in a study of 79 cases of DLBCL of primary cutaneous resulted 3 cases on the chest, 2 cases on the shoulder, 2 cases on the back, 1 case on the trunk, and others on the extremities (9). Another case of shoulder lymphoma has been found in a patient who had undergone shoulder arthroplasty after prolonged immunosuppressive therapy taken because affected by long-standing rheumatoid arthritis (10). A case of DLBCL of shoulder is reported in a 34-year-old lady, also treated for HIV infection (11). In the few reported cases of DLBCL of the chest wall, surgery provided a relatively satisfactory outcome. Luh *et al* (12), reported a patient with DLBCL developing from a long-standing pyothorax of the left lower chest wall. In that case, the patient remained free of local recurrence or metastasis at nine months after surgical resection, without further chemotherapy (13). In Hsu *et al.*'s series (14), 3 of the 4 patients with isolated chest wall lymphoma were managed with surgical resection and adjuvant chemotherapy. In our case the first management of this condition was surgery and after surgery, the patient started chemotherapy CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisone, rituximab), but the mass was excessive. Our patient's staging included an abdominal and brain CT scan, which gave no indication of metastasis. Surgery in soft tissue lymphoma is still controversial. Damron *et al.* (15) are convinced that lymphoma is a non-surgical disease in which chemotherapy and/or radiotherapy are adequate therapeutic strategies and they do not recommend the surgical excision since it would remove a clinical barometer of responsiveness to medical treatment. Biopsy should be only performed to confirm the nature of the lesion, especially in differential diagnosis with soft tissue sarcoma. On the contrary Bozas *et al* (16), described a case of abdominal wall mass (10×18cm) situated between the abdominal muscles and in which a wide excision was performed followed by immuno-chemotherapy. Belaabidia *et al.* (17), also described a

case of muscle lymphoma of biceps femoris (17×14×7cm) in which treatment was wide surgery followed by chemotherapy. In our patient we have applied surgery as a palliative treatment.

Conclusion

A painful shoulder may be caused not only from sarcoma, but also from lymphoma. In suspected patients, plain radiographs should be followed by magnetic resonance imaging and biopsy to determine the right diagnosis and treatment. The malignancy's stage and histopathological diagnosis have a major impact on its treatment and prognosis. Patients with lymphoma are usually treated with chemotherapy. This report does not allow to draw any conclusion about the best treatment and long term outcomes of DLBCL patients. Larger studies with long-term follow-ups are needed to better define the evolution of this pathology.

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